

Case Report
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Hidradenitis Suppurativa: “A surgical Update with Review of Literature”

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ABSTRACT

Hidradenitis suppurativa (HS) is a chronic skin disease primarily affecting the sweat glands apocrine zone of the body and what starts initially as an abscess progresses to nodules, sinus tracts, with cicatrisation fistula formation and ulcers. It is common to see all the stages of clinical spectrum viz multiple solitary abscess to nodules with sinus, fistulas with ulceration and cicatrisation in chronic patients. This chronic illness takes a toll on the psyche of these individuals as the need to avoid foul smell and the constant need for dressing dictate their social life. The patients are notoriously know for follow up for years. A low self-esteem and depression are companions of hidradenitis suppurativa (HS) individuals. Early involvement of the surgeon can substantially improve the progress and evolution of this chronic skin condition and could obviate the development of SCC the end stage of all Chronic neglected Hidradenitis suppurativa (HS) cases especially in the hi risk older age group of individuals.

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Introduction

Hidradenitis suppurativa is the culmination of the clinical spectra of abscesses, sinuses and fistula tracts in evolution and are confined to the apocrine skin fold zones of the body [1]. It is a chronic skin condition that is accurately diagnosed by Clinical examination alone. The patient is often juggled between a community physician, a gynaecologist and a dermatologist before being referred for an adequate drainage technique. Thus there is a delay in the diagnosis of this skin condition [4]. A delay in diagnosis delays appropriate treatment and the chronicity of the disease process implies patients are under a long follow up for years. The cause of this debilitating condition is multifactorial with both hereditary and environmental factors contributing to its development. The initial event in the pathophysiology involves overproduction and subsequent occlusion of the common channel draining hair follicle and sebum unit. This is followed by increased intraluminal pressure and hair follicular rupture, with spill into the subcutaneous tissue. The cellular spill serves as a nidus for bacteria and chronic inflammation orchestrates immune responses that are responsible for chronic inflammation and scarring seen in the skin folds of the affected axillary, genital or perianal regions.

Case Presentation

A 30 year-old Middle eastern obese male (BMI) of 35 kg/m² presented in the clinic with a history of gradually progressive right sided axillary swelling and intermittent lower groin pain with foul smelling discharge at the axillary and later on in the groin wounds as well. This nodular swelling and subsequent fistula and scarring of his wounds had been present for more than eight years. This patient was a staff at Red Crescent Society. After a long follow up with the community physician and dermatology clinic he was referred to the surgical team for opinion.

A routine lab test showed an increase in WBC and CRP, His MRSA swab was negative, all other investigations were normal. O/E middle aged male, who had bilateral axillary grade III Hurley lesions in axillary and bilateral grade II Hurley groin inguinal lesions. He had undergone a wide excision for the grade III Hurley axillary regions and had an uneventful recovery in the post op period. Currently he is on follow up in the clinic and his wounds have healed.



Figure 1A

Figure 1B



Figure 1C

Figure 1A, Right Axillary Grade III Hidradenitis, Fig.1B, Left Axilla Grade III, Fig.1C Right Groin Grade II Hidradenitis.

Discussion

Hidradenitis suppurativa (Greek) when translated it refers to sweat glands that are infected. It also goes by the names Verneuil's disease, Velpeau's disease after the French surgeon who described it. It is a chronic inflammatory condition affecting skin regions bearing apocrine glands zones, the skin folds at axilla, inguinal, perianal and the inframammary regions. It is a disease condition affecting the hair follicles [28].

The prevalence of hidradenitis suppurativa in the studies in Europe is reported at 1% in the general population the incidence is higher in females compared to males 3: 1 [2,3,25]. Cigarette smokers are at increased risk of acquiring hidradenitis suppurativa [3,37]. Hidradenitis suppurativa dwells with the overweight and obese individuals. The most common clinical presentation is pain, swelling with pruritus followed by the development of a skin nodule over a few days [3]. A mechanical skin friction in the apocrine skin fold zone opens up micro ulcers, this coupled with an inherited dysregulated system response of macrophages in visceral fat that secrete increased levels of cytokines flares up disease activity [15]. There appears to be a familial component as 30%–40% of Hidradenitis suppurativa patients reported a family history of Hidradenitis suppurativa in relatives [3,5]. The familial forms of hidradenitis suppurativa follow an autosomal dominant pattern of inheritance. The mutations in subunits of the proteins is seen in 5% of hidradenitis suppurativa cases [6,7]. Hidradenitis suppurativa is also prevalent in the paediatric obese children, it is common in the age group of eleven years. As with the adult hidradenitis suppurativa it is more common among girls than boys. There is a hereditary component to the paediatric hidradenitis suppurativa, similar to its adult counterpart.

The pathophysiology of Hidradenitis suppurativa involves increased production of keratin cells within the hair follicle followed by occlusion at the common draining the sebum, hair follicle channel and subsequent dilatation. With progressive increased intraluminal pressure there is a rupture and spill into surrounding dermis that triggers a vigorous cellular response from neutrophils and lymphocytes. The inflammatory cellular infiltrate causes a localised abscess formation, and a contracture of the subdermal tissue with subsequent fibrosis. Gross axillary pads of fat in Obese individuals provide mechanical friction at the skin folds this epithelial invagination coupled with excess apocrine secretion form the perfect nidus for bacterial implantation and infection. The Epidermal invaginations produce chronic discharging sinus tracts from abscesses. The sinuses fistulate with fibrosis of the dermal tissues and ulcer formation. The most frequent organism to be isolated is *Staphylococcus lugdunensis* that is present in more than 50% of Hidradenitis nodules and other organisms include Streptococci, anaerobes and E coli [13,36].

The reason for chronic inflammation is the formation of bacterial biofilms that bind irreversibly to sinus tract epithelium and hair follicles thus inciting a chronic inflammatory response [17]. It is this persistent chronic inflammatory phase that is a trigger for an immune response. A dysregulation of innate and adaptive immune is then responsible for the full spectrum of nodular abscesses with chronic discharging sinuses and fistulas seen in grade two and three Hurley hidradenitis suppurativa [12]. Melnik and Plewig have proposed the concept of a self-inflammatory disease characterized by dysregulation of the Notch pathway [8,9]. Notch signalling is of

prime importance for maintaining the inner and outer root sheath of the hair follicle appendageal cellular harmony and integrity. Any deficiency in this pathway results in compromise of the apocrine gland homeostasis, that triggers stimulation of innate immunity, that supports and maintaining chronic auto inflammatory state [10,11].

The Hurley Clinical staging system classifies Hidradenitis into three stages [14].

- **Grade I:** Abscess formation, single or multiple, no sinus tracts, no cicatrization
- **Grade II:** Recurrent abscesses along with formation of tracts and cicatrization either solitary or multiple.
- **Grade III:** Diffuse skin involvement, and organisation of multiple interconnected abscesses and tracts.

As with any persistent chronic inflammatory state the risk of transformation to squamous cell carcinoma (SCC) is 1% to 3.1% (16). This risk in another cohort study was reported at a slightly higher rate of 4.6% [27]. The SCC arising in chronic inflammatory disorders carry a high mortality of 40%. A surgical consultation is imperative in those high risk groups older aged individuals with chronic discharging fistulation of wounds. The risk of SCC transformation is higher at extra axillary sites like groin and perineum in comparison to axillary hidradenitis suppurativa.

Differential diagnosis of hidradenitis suppurativa includes carbuncles, inflamed pilonidal cysts, lymphadenitis, fistula producing diseases like tuberculosis, actinomycosis, lymphogranuloma venereum, inguinal lymphatic granulomatosis and ulcerative colitis.

Treatment of hidradenitis suppurativa in Pediatric and Adults

General measures are offered for all patients and include weight loss and tobacco abstinence. The goals of therapy are disease control and management of pain. Hurley grade I, Mild cases can be treated with topical antibiotics. Clindamycin is the only antibiotic that has been studied as a topical agent for hidradenitis suppurativa grade I superficial lesions folliculitis, papules, and pustules A dual combination therapy of drugs is recommended for deeper nodules and abscesses Hurley grade II and early grade III [3,19].

Modalities like biologic agents, surgery laser therapy are reserved for severe grade III cases of hidradenitis suppurativa. A useful algorithm for the management of hidradenitis suppurativa in adults, is the Grading of Recommendations Assessment and Evaluation (GRADE) methodology that is an Evidence based approach for making recommendations in clinical practice [32].

Dual modality therapy

Systemic treatment clindamycin + rifampin/tetracycline/acitretin as treatment for Hurley II stage (18). Retinoids are synthetic aromatic derivatives of vitamin A that regulate cell proliferation and differentiation of keratinocytes and restrict inflammation. The dose of isotretinoin is 1 mg/kg/day as monotherapy. Retinoids may also be useful only as an adjunct to reduce inflammation before and after surgery [29].

Molecular therapy

Systemic biologics (adalimumab/infliximab) are reserved for treatment of resistant, moderate to severe hidradenitis suppurativa (moderate Hurley II/Hurley III stage) the combined use of systemic 300 mg of clindamycin twice a day and rifampicin 600 mg daily given as either 1 or 2 doses for 10 weeks is beneficial, with 47%

of remission in patients and others reporting a total remission after 10 weeks [20,21].

Adalimumab and infliximab, two different monoclonal antibodies against TNF- α , are effective in the treatment of moderate to severe HS (Hurley II–III) [22,23]. Infliximab (5 mg/kg body weight) is administered intravenously over a period of 2 hours on day 0, 2, 6, and then regularly every 8 weeks Adalimumab, a human monoclonal antibody that binds to and neutralizes TNF- α , (RCT trial PIONEER I and PIONEER II) It is administered adalimumab at a weekly dose for 24 weeks [24].

Surgical Modality as Treatment

Surgical intervention is the modality of choice for recalcitrant lesions. Those with diffuse scarring and rope like extensions require wide excision with or without skin grafts or rotation and advancement flaps [30]. A mere drainage procedure is not enough for inflamed lesions with nonfluctuating nodules. The rate of recurrence is very common if the chronic recurrent nodules are merely incised for localized collections. Surgical de roofing of the sinus tract leaving the wound for secondary healing is a temporary option. Wide excision with lateral margins from 5 mm up to 2 cm however, of all hair bearing appendageal structures in the affected region gives better cosmetic result [21,26]. In contrast, a wide local excision and deep margin to fascia and is a curative method of management. Any potential fistulas must be identified and excised, using injecting 3-5 mL of a methyl-violet solution [33]. The rate of recurrence is 3% for axilla, 0% for perianal region, 37% for inguinoperineal and 50% for sub mammary regions after the follow-up from 3 to 72 months [34,35].

Skin Tissue Sparing Excision using Electrosurgical Peeling Procedure (STEEP)

STEEP is done with the patient under general anaesthesia. The goal of this procedure is to achieve complete removal of affected tissue and spare as much healthy tissue as possible to prevent the formation of contractures. The principle is the use of successive tangential trans sections that allows deep excision of fibrotic and inflammatory tissue thereby leaving the epithelialized bottoms of the sinus tracts and a large pad of the subcutaneous fat intact the end result is a cosmetic smaller defect. STEEP electrosurgery is reported to have important advantages over the carbon dioxide laser, it is more easily controlled and adjusted by the surgeon. The carbon dioxide laser on the other hand removes a continuous horizontal plane of one depth, making it less precise and less tissue sparing [36].

Conclusion

Hidradenitis suppurativa is a chronic debilitating condition. The clinical spectrum of chronic abscesses are in a state of dramatic evolution into sinuses, fistulas and chronic ulcers that has a devastating impact on the psyche of the individual. An early involvement of a General Surgeon can avert the potential development of SCC especially in high risk individuals. STEEP electrosurgery is an innovative technique, only further studies will determine its definite role in the management of Hidradenitis suppurativa.

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